

New ophthalmology course

AFCM

Problems of external appearance
II. Orbital diseases and Proptosis

INTENDED LEARNING OBJECTIVES

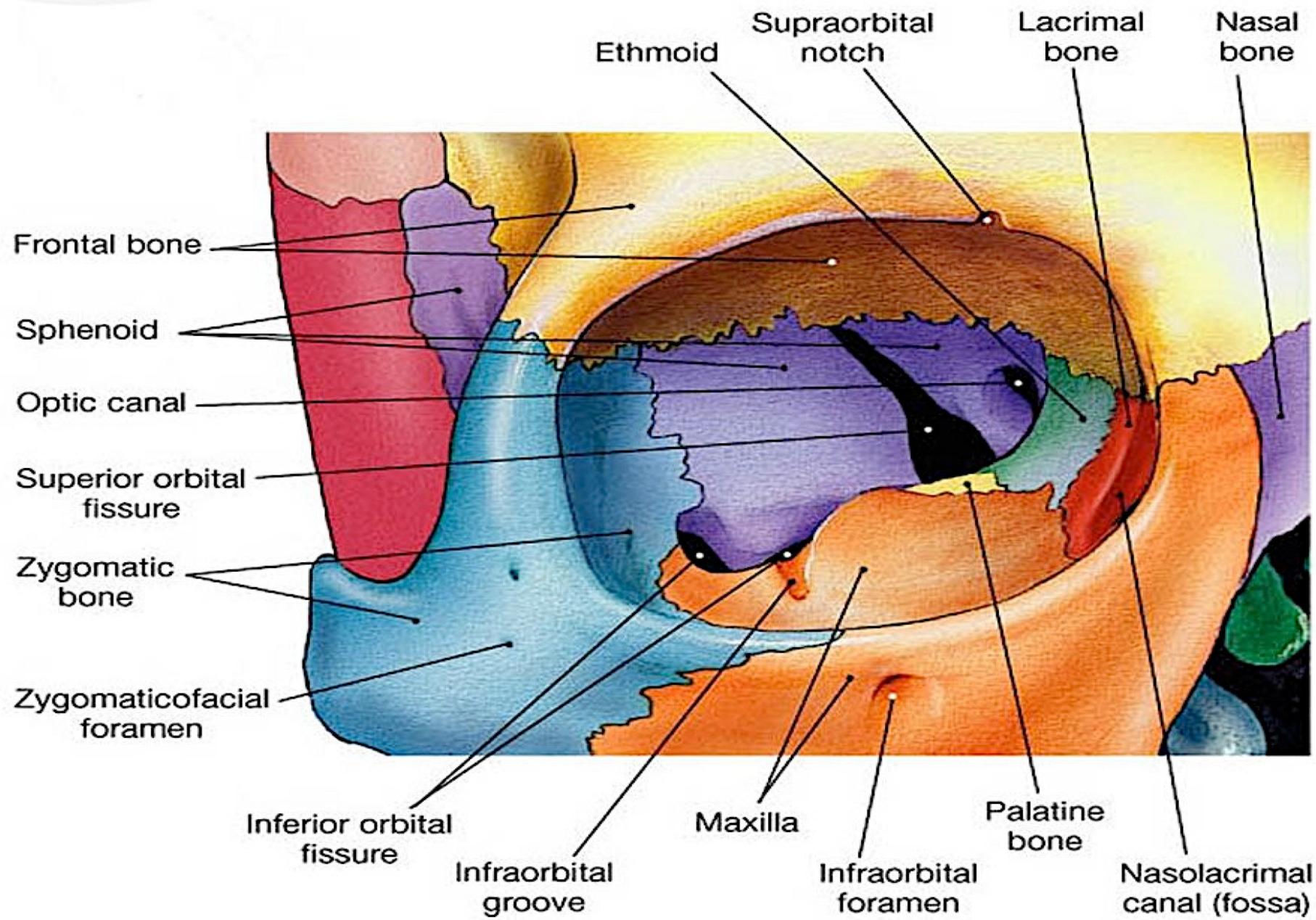


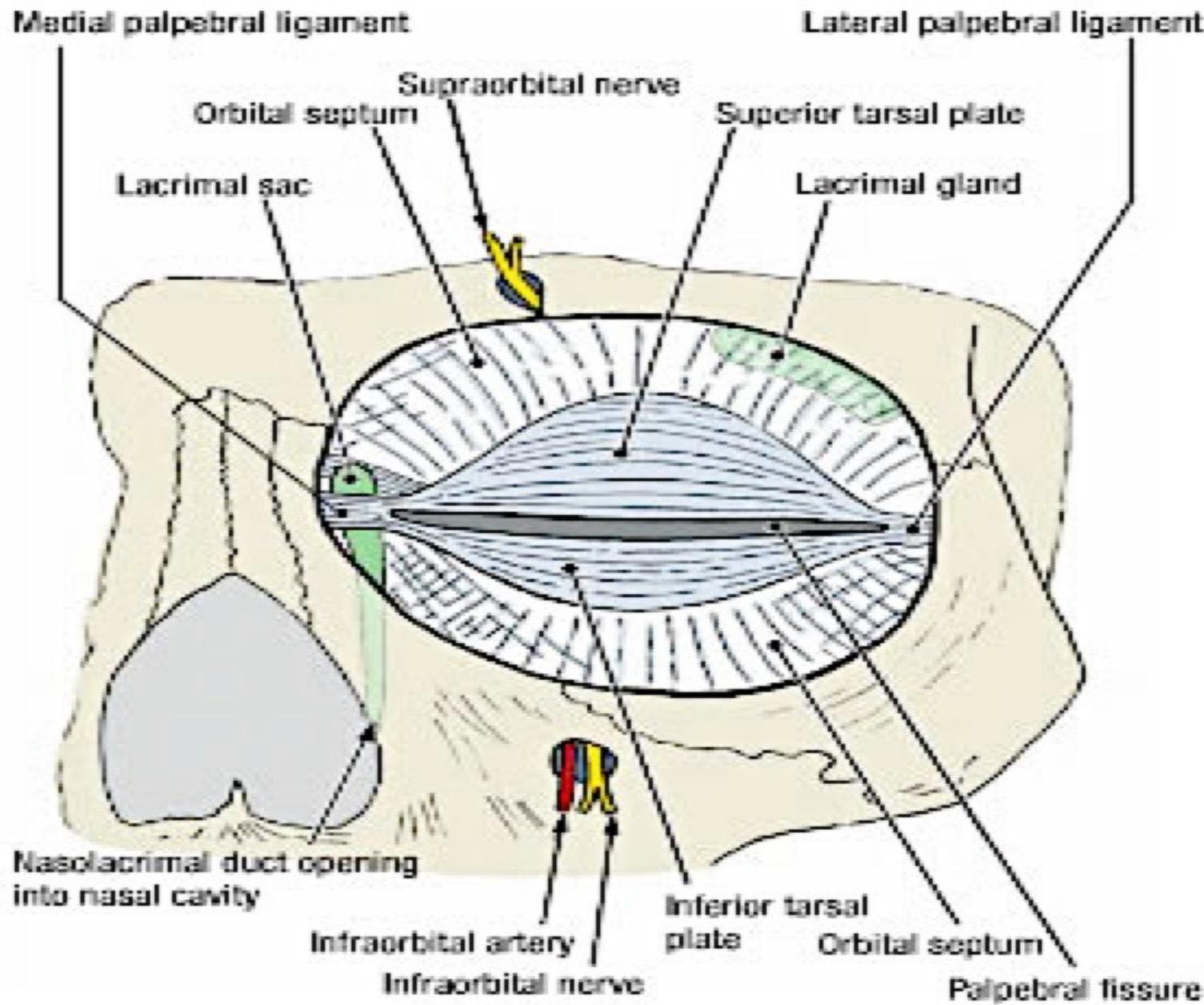
By the end of this lecture the student will be able to:

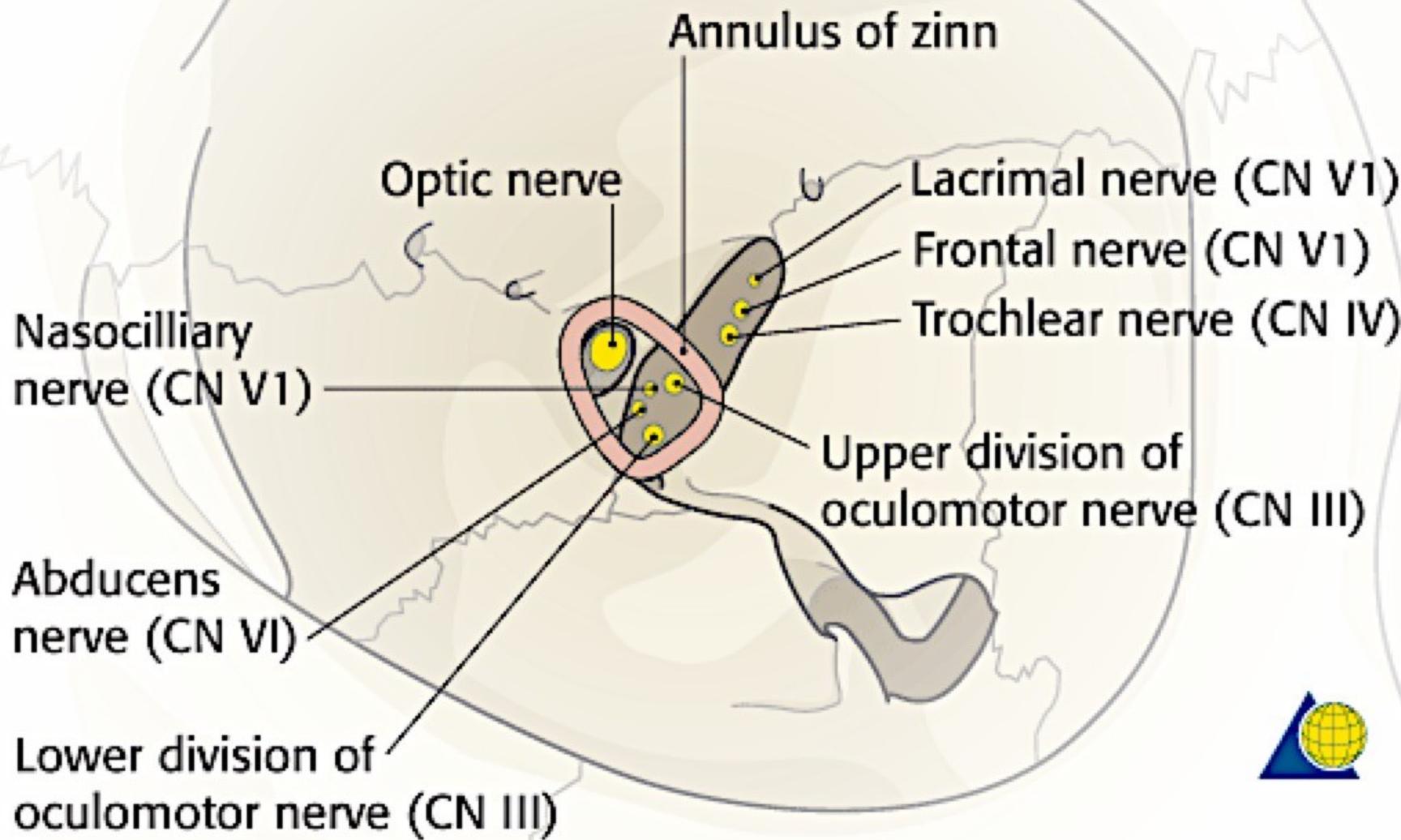
1. Describe the main features of orbital anatomy
2. Understand the techniques of orbital examination
3. Know the common orbital diseases
4. Understand the principles of orbital investigations
5. Recognize the features of TED
6. Evaluate the effects of thyroid eye disease on the eye and orbit
7. Solve clinical problems in orbital disease

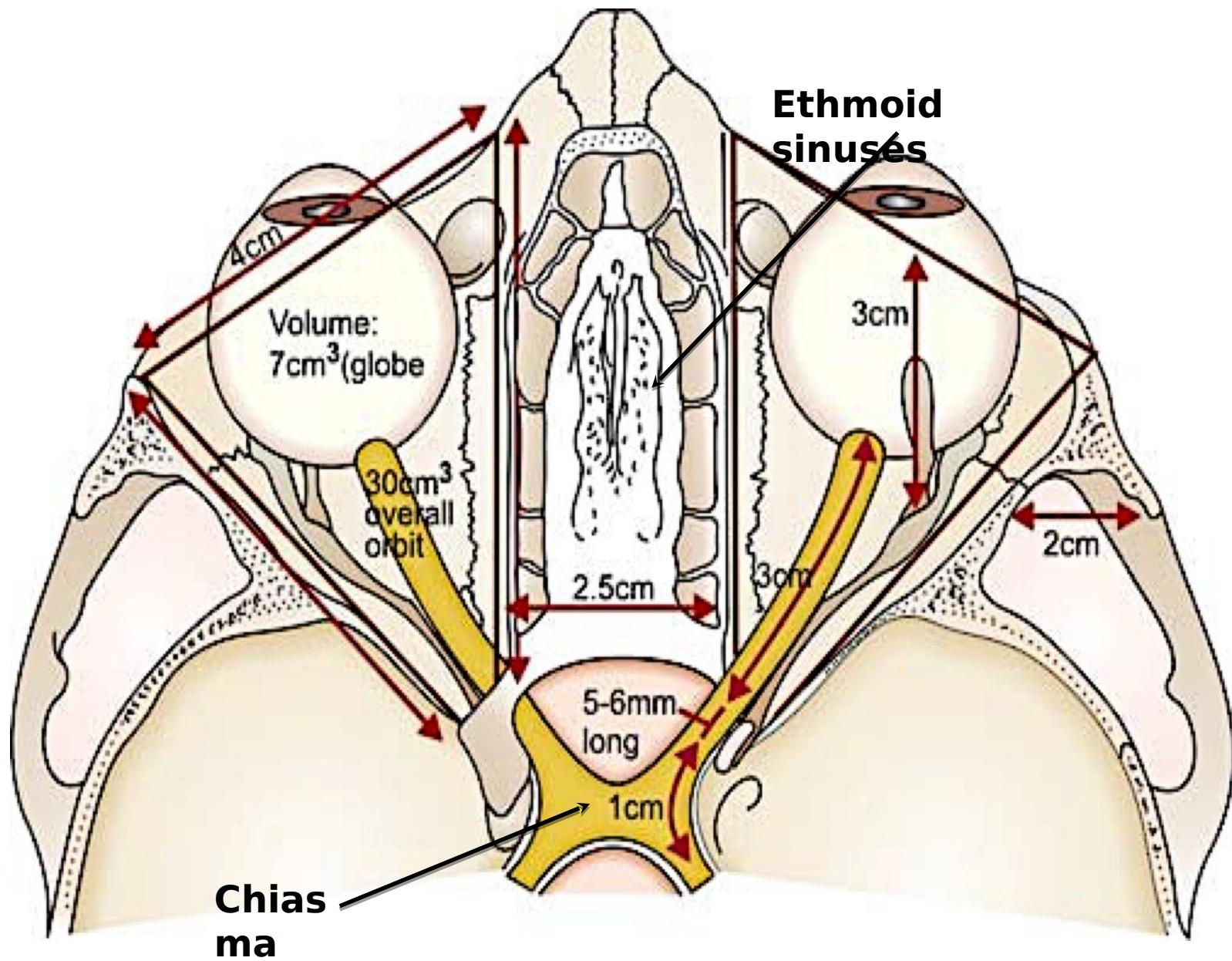
Orbital diseases

- Anatomy
- Examination
- Exophthalmometry
- Proptosis
 - Children
 - Adults
- Inflammation
 - Orbital cellulitis
 - TED
- Trauma
- Investigation
 - CT scan
 - MRI









Proptosis



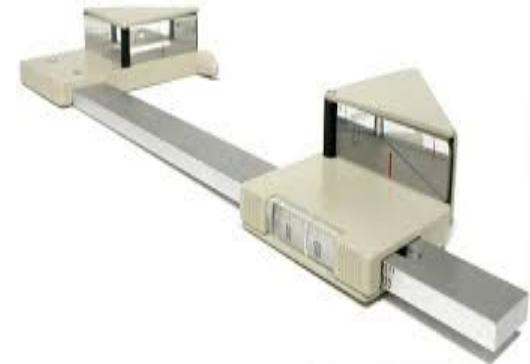
- Forward Protrusion of the eye ball



How to examine a case of proptosis?

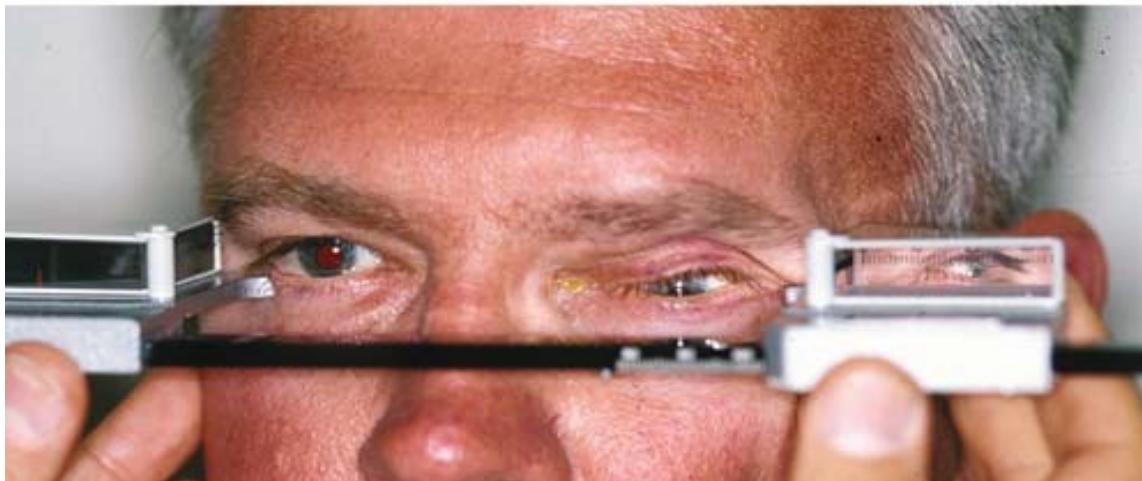
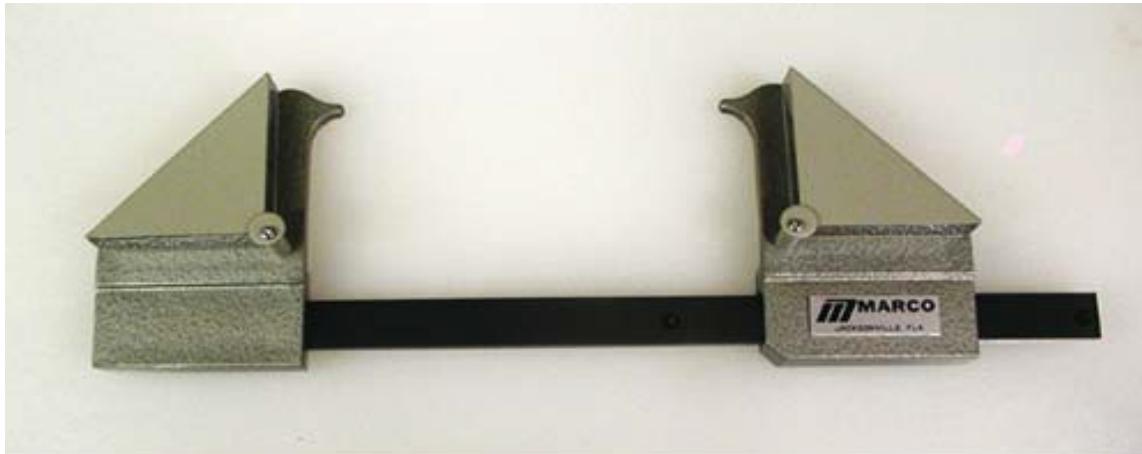


- Assessment of proptosis
 - **Identify**
 - **Direction**
 - **Measure**



- IDENTIFY

- Lid positions
- Large globe (pseudoproptosis)
- Shallow orbits
- Globe displacements
- Old photos



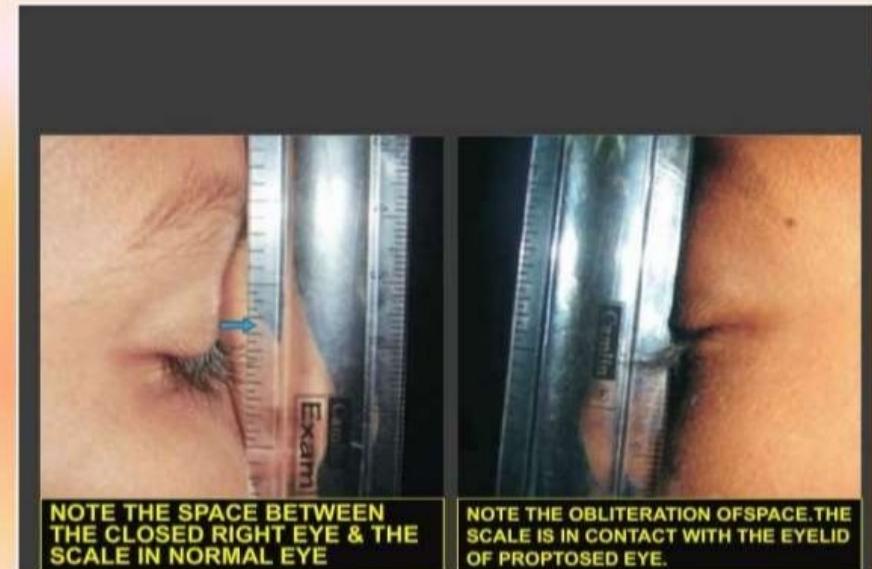
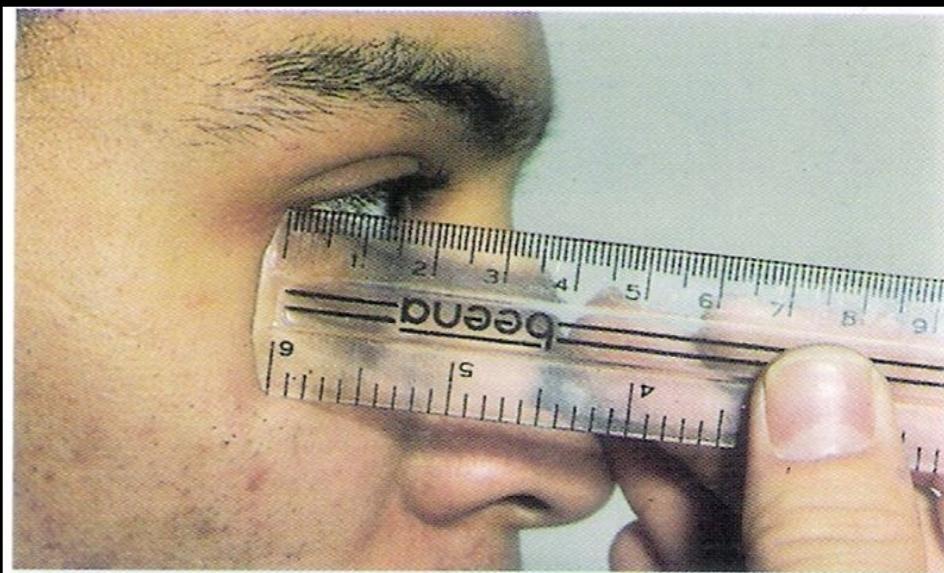
**Hertel's
exophthalmometer
(MEASURE)**

- Proptosis is measured from the corneal apex to the **margin** of the zygomatic arch.
- White men =17-21 mm and 16-20 mm in white women.
- Black adults, measurements are increased by 2 mm
- An exophthalmometry measurement >21 mm or a difference >2 mm between the two eyes is usually considered abnormal.
- Enophthalmos measurement **<14 mm**

Inspection from below

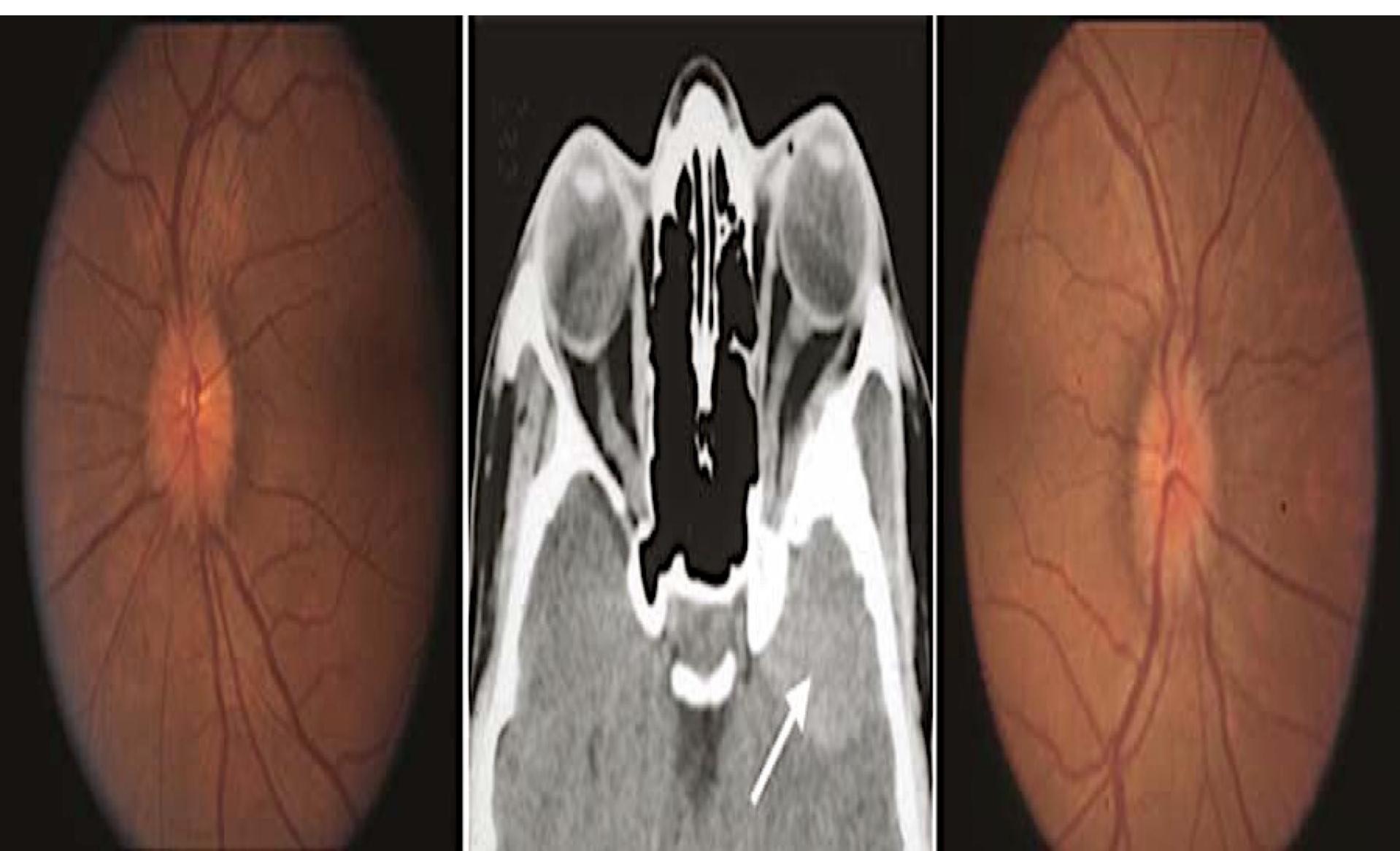


Clinical test; measuring proptosis



The ocular examination should include:

1. Best-corrected visual acuity
2. Intraocular pressure applanation
3. Extraocular muscles motility
4. Fundus examination
5. A neuro-ophthalmologic examination
 - a. color vision assessment
 - b. contrast sensitivity
 - c. pupillary examination
 - d. motor and sensory functions
 - e. confrontation visual fields and central visual field with an Amsler grid



Papilledema in a case of sphenoidal ridge
meningioma

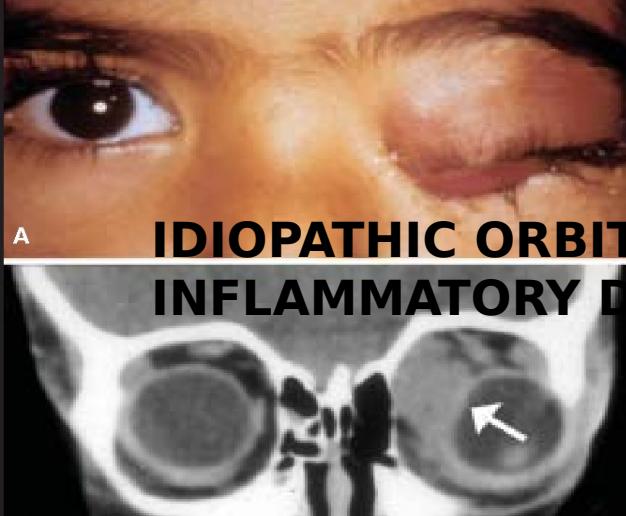
Investigation of Orbital Disease

- Thyroid function tests
- Complete blood picture
- ESR, CRP
- CT scan/MRI
- Fine Needle Aspiration Biopsy (FNAB)
- Surgical biopsy

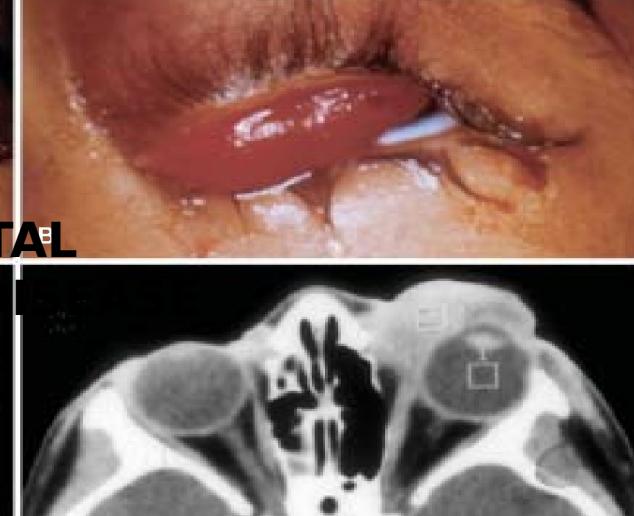
Common causes of proptosis in children

- Unilateral
 - Acute/Inflammatory
 - **Orbital cellulitis**
 - Idiopathic inflammatory orbital disease
 - Rhabdomyosarcoma
 - **Orbital extension of retinoblastoma**
 - Benign tumors
 - Capillary hemangioma
 - **Lymphangioma**
 - Dermoid cyst
 - Optic nerve glioma
 - Optic nerve meningioma
- Bilateral
 - Craniosynostosis
 - Metastatic (neuroblastoma, leukemia, reticuloendothelioses)
 - Idiopathic orbital inflammatory disease **OID**
 - Thyroid eye disease **TED**

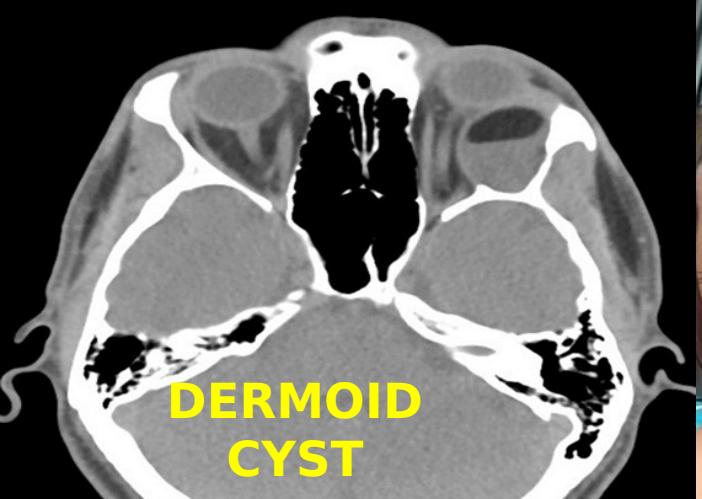
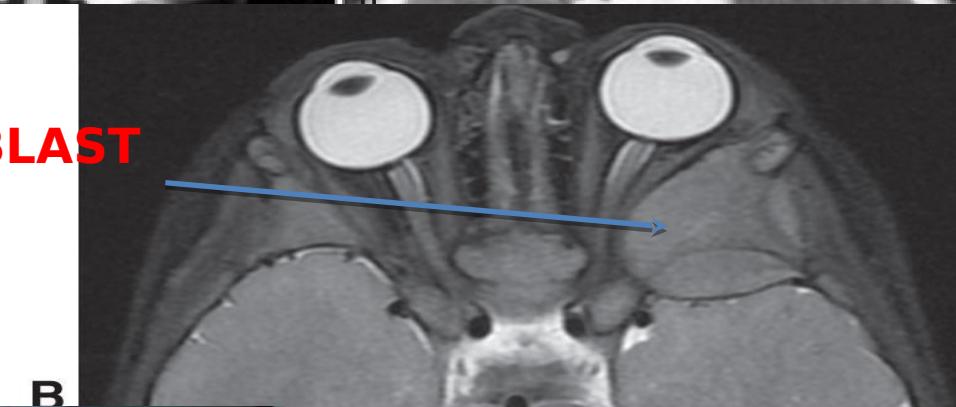
EXTENSION OF
RETINOBLASTOMA



IDIOPATHIC ORBITAL
INFLAMMATORY D



NEUROBLAST
OMA



DERMOID
CYST



Crouzon
Syndrome



Acute
leukemia

Common causes of proptosis in adults

- Unilateral
 - INFLAMMATORY
 - Thyroid eye disease
 - Orbital cellulitis
 - Idiopathic orbital inflammatory disease
 - Benign tumors: cavernous hemangioma, Schwannoma, dermoid cyst, **MUCOCELE**
 - Malignant tumors AND **SECONDARIES**
 - Optic nerve tumors
 - Carotid cavernous fistula

Common causes of proptosis in adults

- Bilateral
 - INFLAMMATORY
 - Thyroid eye disease (TAO, TED)
 - Idiopathic orbital inflammatory disease (IOID)
 - Cavernous sinus thrombosis
 - Lymphoma
 - Bilateral lacrimal gland swelling
 - Neurofibromatosis type I

Idiopathic Inflammatory Orbital Pseudotumor

- History: Acute, recurrent, chronic. Any age
- Clinical features: **pain**, redness, lid swelling, diplopia, proptosis, decreased ocular motility
- Types:
 - **orbital myositis**
 - apical idiopathic orbital inflammation
 - sclerosing inflammation
 - lacrimal gland: granulomatous, vasculitic
- Testing:
 - Examination and vitals
 - Ultrasound: irregular, uniform density lesion with
 - ESR, CRP, CBC, IgG4, ANCA
 - **MRI**
 - Orbital biopsy
- Treatment:
 - **Steroids**



Orbital Cellulitis



- Bacterial Inflammation of the soft tissue behind the orbital septum



Orbital Cellulitis

Etiology

- Extension from sinus infection: 90%, ethmoidal sinus: 90%
- Orbital trauma/surgery
- Organisms: staph, strep, H-flu, and gram - rods

Clinical picture:

Pain, Fever, Proptosis, limitation of EOM motility

Testing:

- Labs: CBC with differential, chemistry panel, culture: blood, wounds, debrided material
- CT/MRI: edema, inflammatory material and cells in sinus/orbit

Orbital Cellulitis

Treatment:

- Admission to hospital: referral to ENT, Internal Medicine, and Ophthalmology
- Nasal decongestant spray , ANTIBIOTIC ointment topically: culture specific
- Surgical drainage of abscesses
- I.V. ANTIBIOTIC x 1 week, response w/in 48 hrs.

Orbital Cellulitis: DD



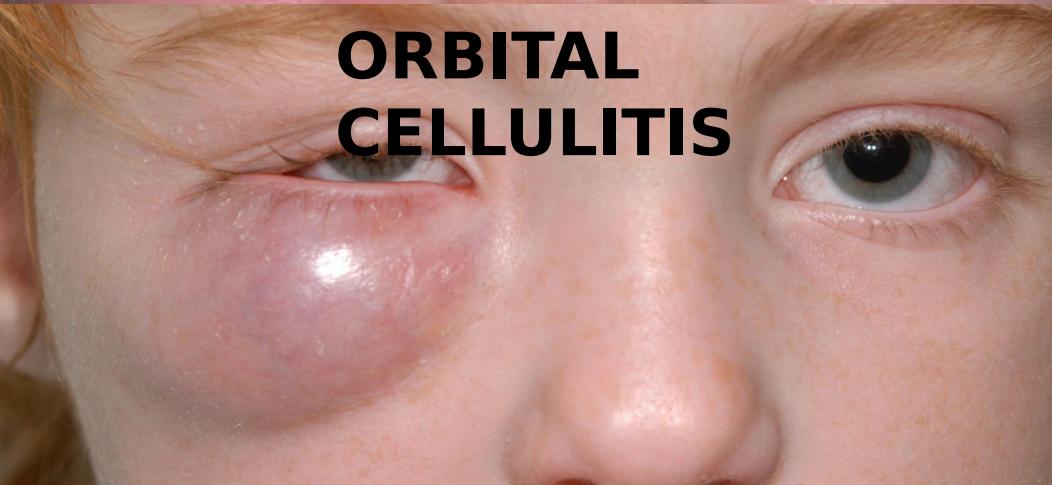
**Rhabdomyosarcoma
Leukemia**



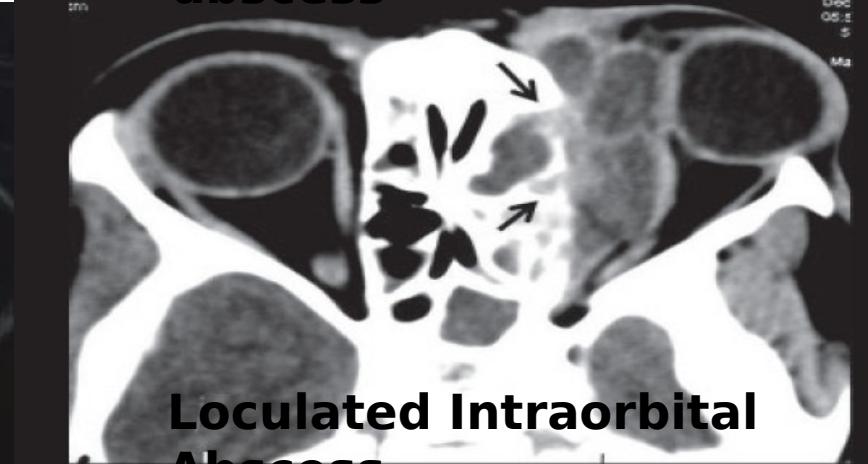
Rhabdomyosarcoma of the orbit.

Brini, Dhermy, Sahel.

Oncology of the Eye and Adnexa, 1990.



**ORBITAL
CELLULITIS**



Thyroid-Associated Ophthalmopathy Thyroid Eye Disease

Thyroid Eye Disease

General

- most common cause of orbital disease
- most common cause of proptosis
- F > M 6:1
- Most common in females 30-50 yrs.
- course of eye involvement does not parallel systemic disease, diagnosis is clinical
- **30%** of patients with immune thyroid diseases develop orbital involvement and 5% develop more severe consequences
- **Cigarette smoking** is strongly correlated with the development of TED
- TED has active phase (6-18 month) then remission/ inactivity
- Management during active phase is aimed at reducing the immune response and inflammation, by **steroids, radiotherapy, or other immunosuppressive agents**

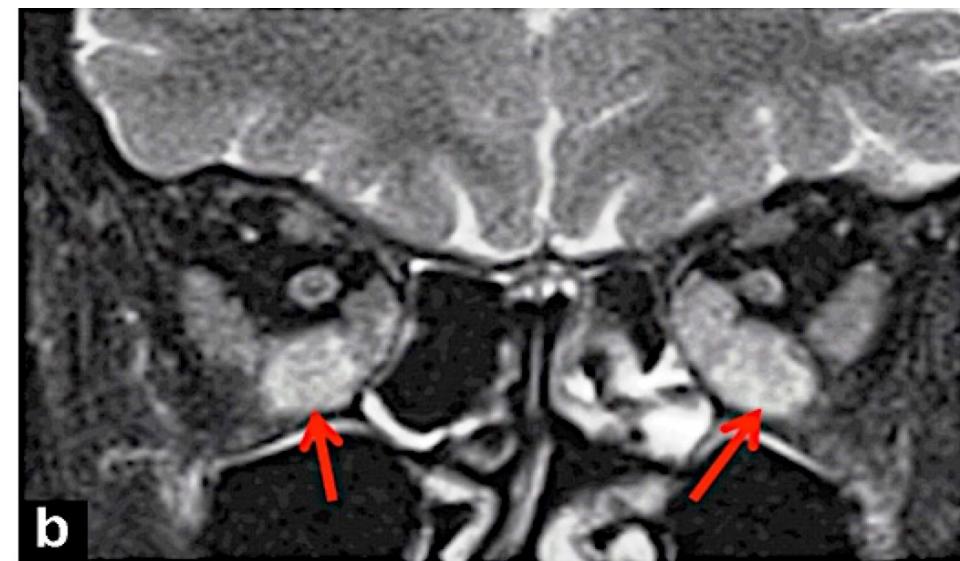
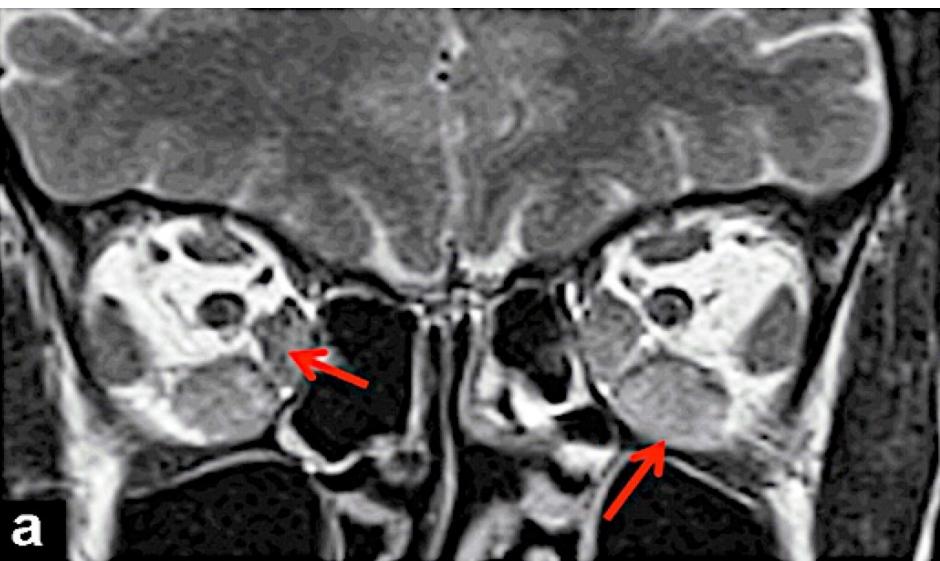
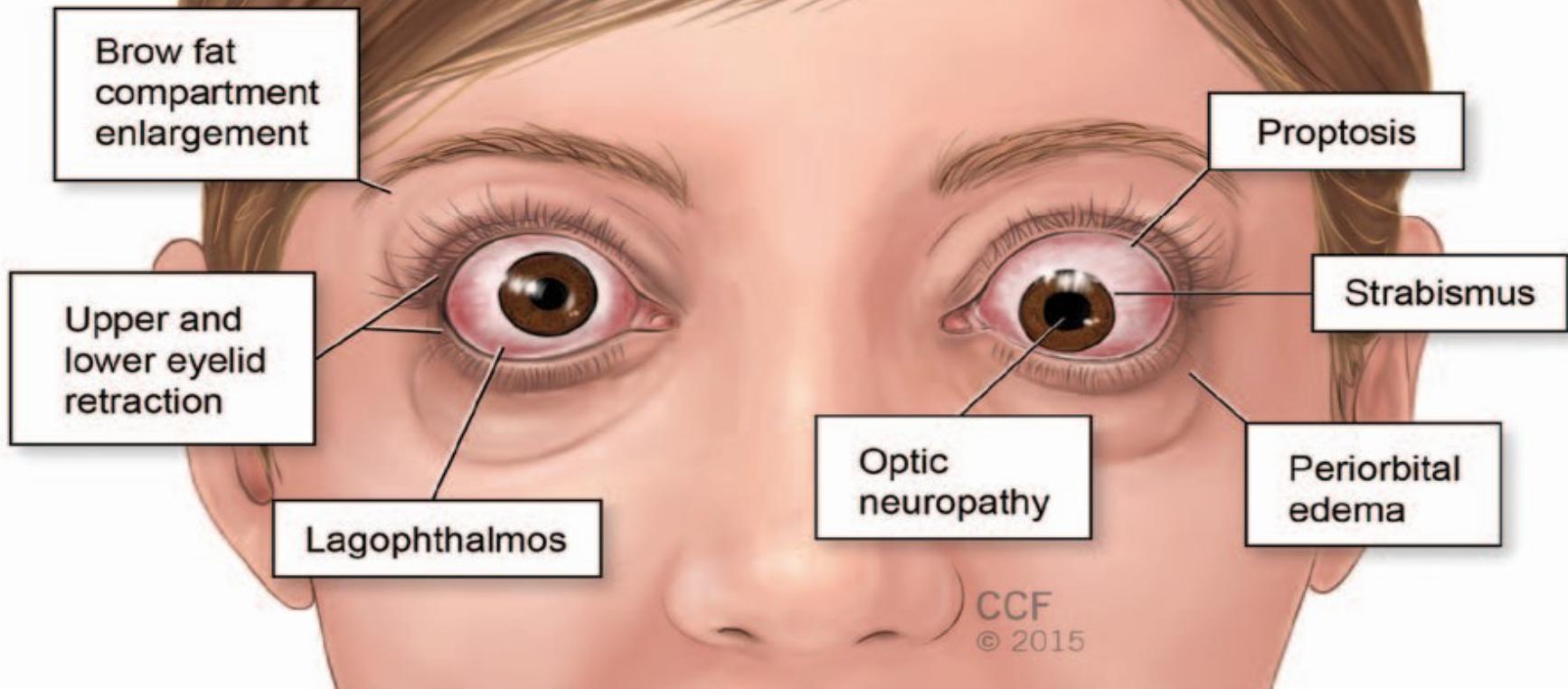
- TED is an autoimmune disease caused by the activation of orbital fibroblasts by autoantibodies directed against thyroid receptors.
- T cell +   ↑ fibroblast 
expression 
Deposition of GAG 
orbital fat expansion

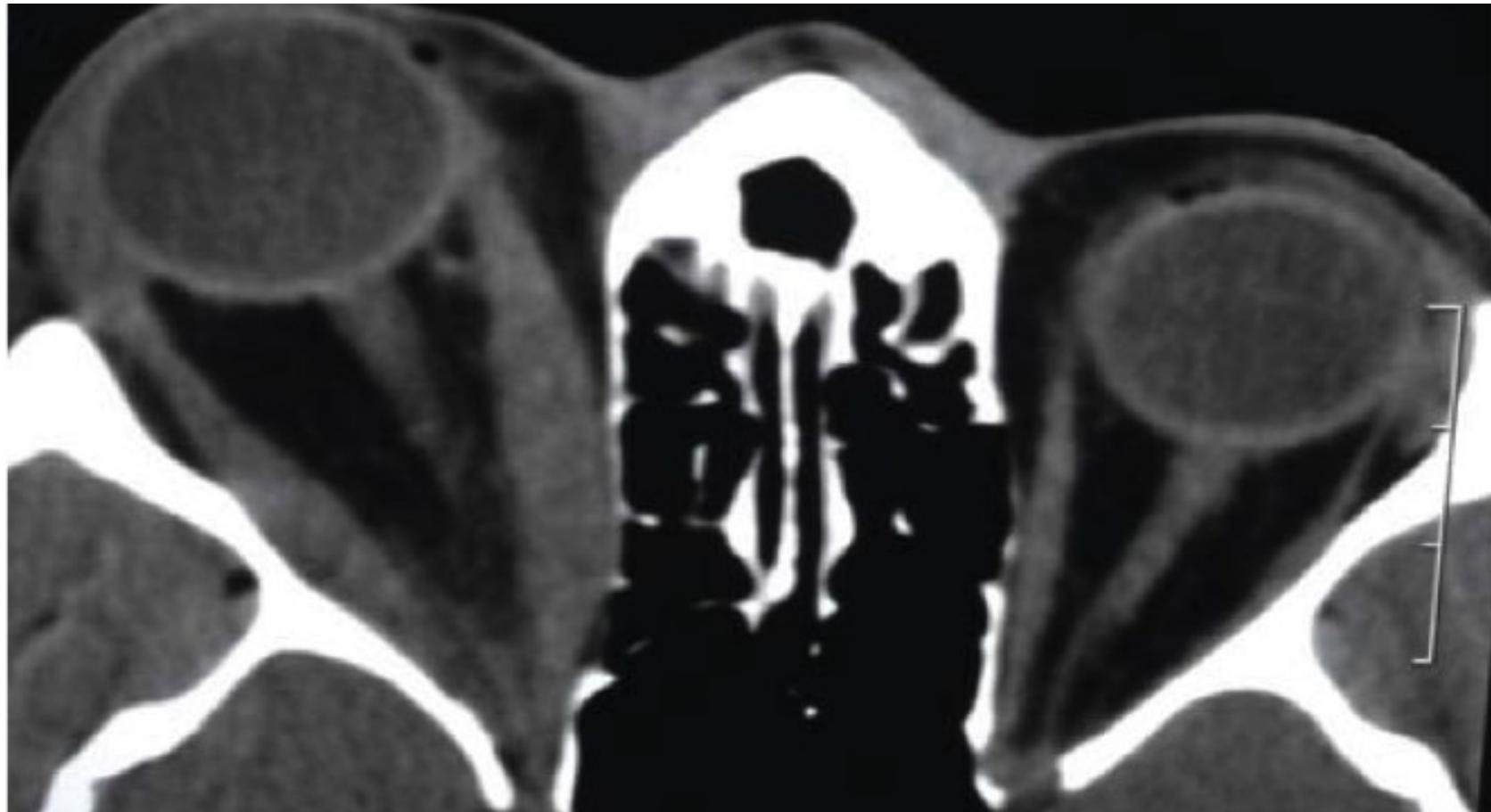
Grave's Clinical Features

- Lid retraction: 90%
- Exophthalmos: 60%
- EOM restriction: 40%
- Injection on EOM insertion: 30%
- Pain and discomfort: 30%
- Diplopia: 17%
- Blurry vision: 7.5%
- Optic neuropathy: 6%

LID SIGNS

1. Dalrymple sign (lid retraction)
2. von Graefe (lid lag)
3. Vigouroux sign (eyelid fullness)
4. Stellwag sign (incomplete and infrequent blinking)
5. Grove sign (resistance to pulling down the upper lid)
6. Joffroy sign (absent creases in the forehead on superior gaze)
7. Möbius sign (poor convergence)
8. Ballet sign (restriction of one or more extraocular muscles)
9. Goldzeiher's sign (injection of the lateral bulbar conjunctiva)





Axial CT scan of patient with asymmetric TAO demonstrating right proptosis from **marked enlargement** of the right orbital muscles and mild enlargement of the fat compartment. Compare the affected right orbit with the normal left orbit

Diagnostic procedures

- **Labs**

T3, T4, thyroid-stimulating immunoglobulins (TSI), anti thyroid antibodies.

- **Ultrasound**

- shows bilateral, asymmetric enlargement of the extraocular muscles. The **muscle belly's are typically enlarged** and the tendon insertions are **spared**.

- **CT Scan**

- Proptosis of globe
 - increased lucency of fat
 - multiple enlarged extraocular muscles (inferior and medial rectus most commonly involved)
 - enlarged superior ophthalmic vein.

- **MRI**

- Fusiform rectus enlargement and orbital fat expansion

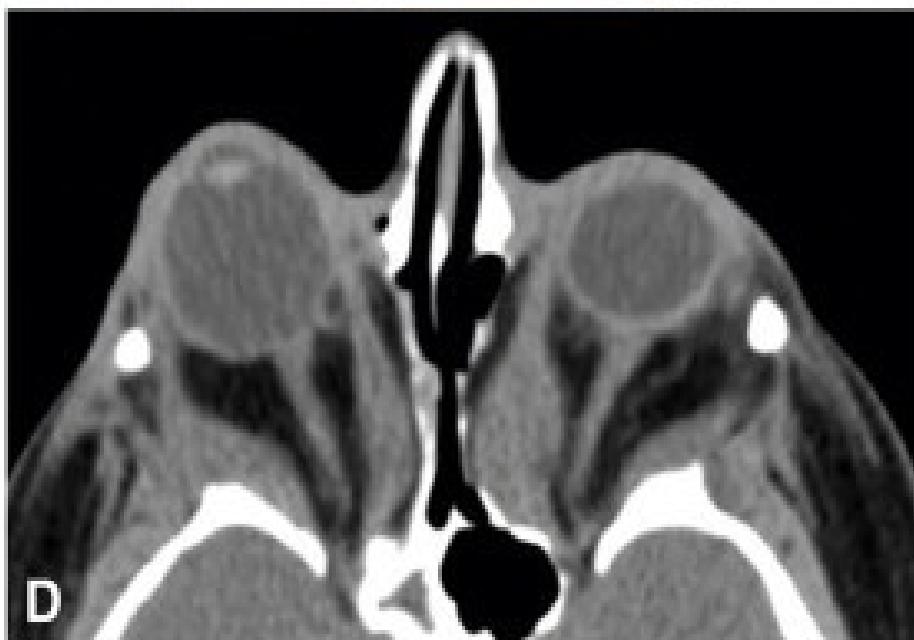
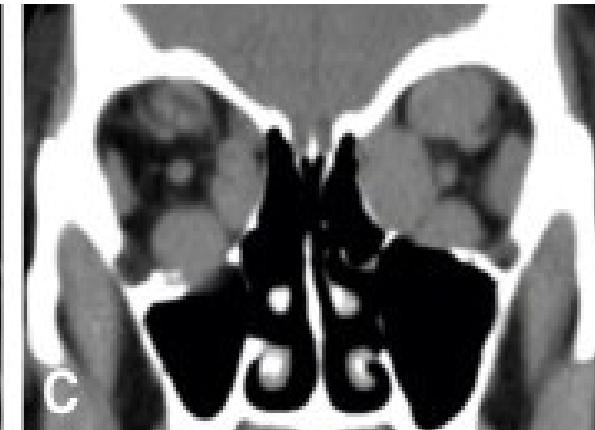
Treatment of TED

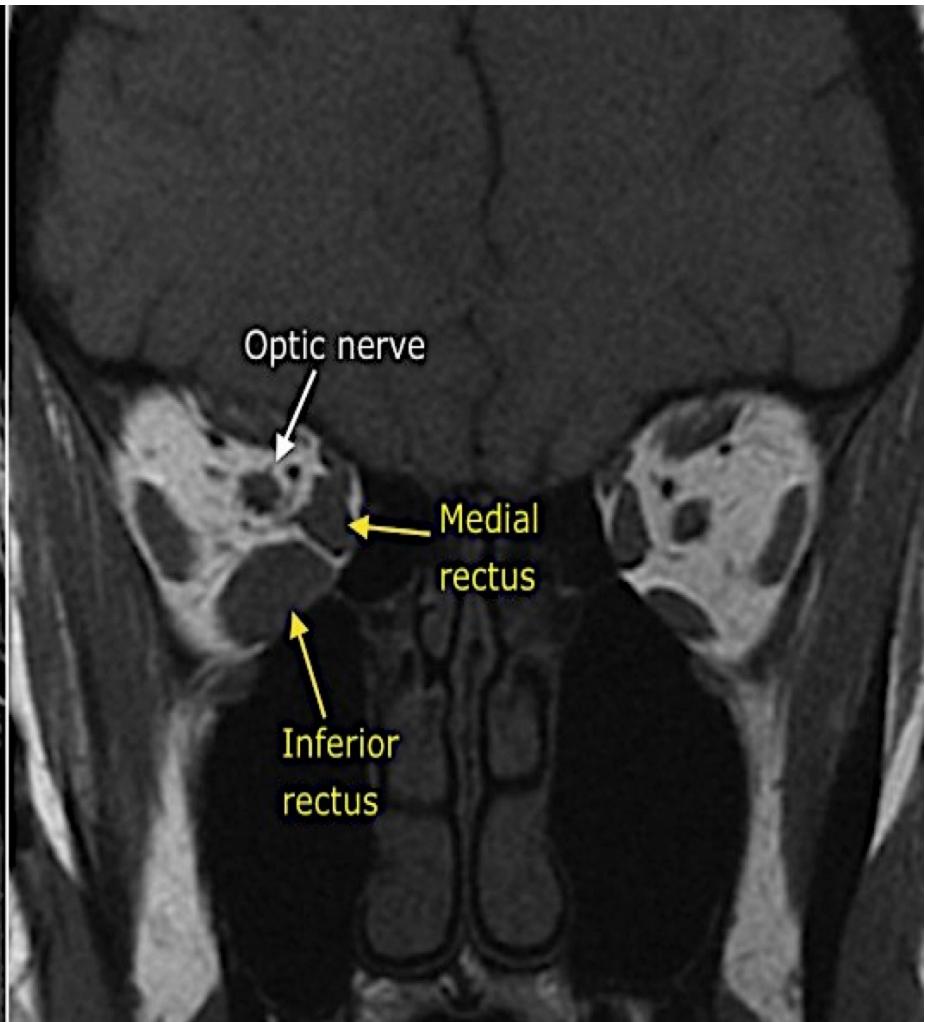
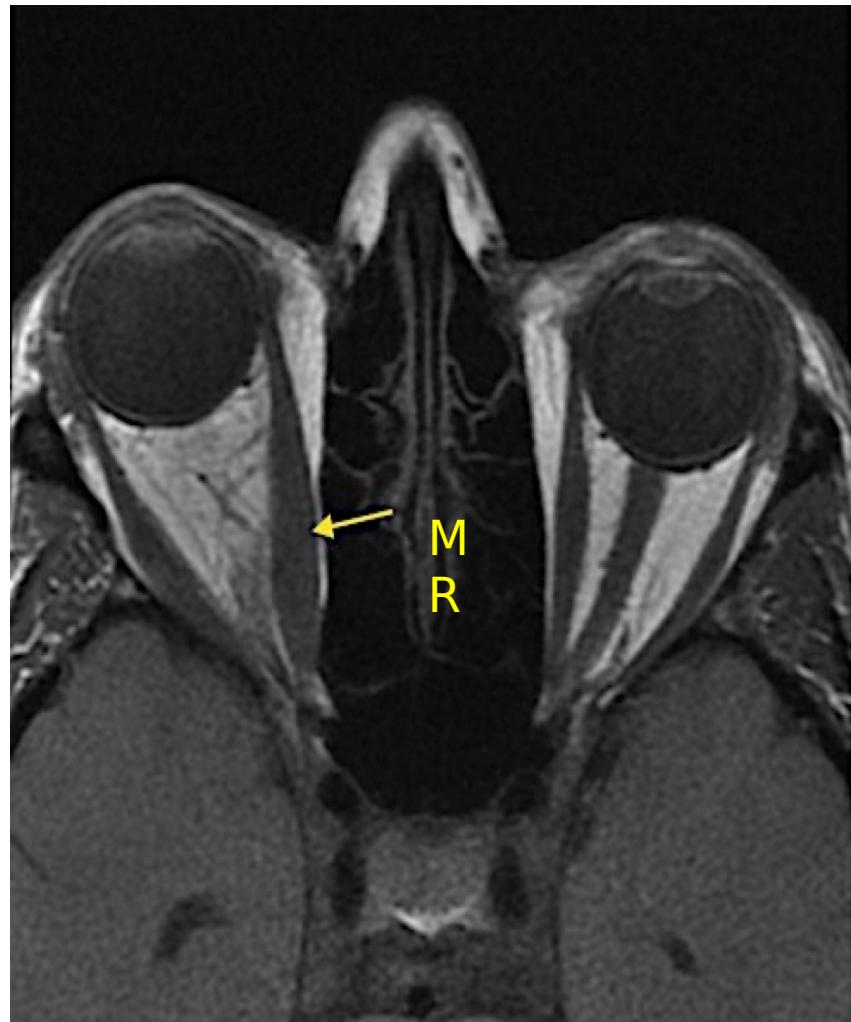
- **Conservative**
 - Stop smoking, oral NSAID, salt restriction, head elevation during sleep
 - Corneal exposure: Lubricants, taping lid, tarsorrhaphy
 - Diplopia: Fresnel prisms, occlusion
- **Biologics:**
 - Teprotumumab
- **Steroids**
 - Oral prednisone in a dose of 1- 1.5-mg/ kg for 2 months.
 - IV pulse methyl prednisolone

Treatment of TED

- Radiation
- Orbital decompression
 - Orbital floor, medial wall, and lateral wall
 - For optic neuropathy and severe proptosis
- Strabismus surgery
 - after orbital decompression and muscle alignment has stabilized.
- Lid retraction repair & tarsorrhaphy

ORBITAL DECOMPRESSION FOR TED OPTIC NEUROPATHY





Orbital diseases should always be referred to higher level care